

Giant solitary synovial osteochondromatosis of the first metatarsophalangeal joint: A case report and literature review.

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ABSTRACT

Giant solitary synovial osteochondromatosis is a rare form of primary synovial osteochondromatosis that exceptionally affects small joints. It often poses a problem of differential diagnosis with chondrosarcoma. We report the observation of a giant synovial osteochondromatosis evolving for many years in the 1st metatarsophalangeal joint

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of the hallux, whose management consisted of an excision of the mass and a definitive arthrodesis given the extent of joint damages.

KEYWORDS: Giant; Osteochondromatosis; Metatarsophalangeal joint; Hallux.

RESUME

L'ostéochondromatose synoviale géante et solitaire est une forme rare des ostéochondromatoses primaire et affecte exceptionnellement les petites articulations. Elle pose souvent le problème de diagnostic différentiel avec le chondrosarcome. Nous rapportons l'observation d'une ostéochondromatose synoviale géante évoluant depuis plusieurs années au niveau de l'articulation métatarso-phalangienne de l'hallus dont la prise en charge a consisté en une résection et arthrodèse vu le constat des dégâts articulaires en per-opératoire.

MOTS-CLÉS : Géant; Ostéochondromatose; articulation métatarso-phalangienne; Hallux.

INTRODUCTION

Giant solitary synovial osteochondromatosis (GSSCM) is a rare form of primary synovial osteochondromatosis (SOC) that exceptionally affects small joints[1]. It causes osseous erosions and destruction of the joints and sometimes dislocations. It often poses a problem of differential diagnosis with chondrosarcoma, especially as there is often hypercellularity in cytological examination, cellular atypies and bi-nuclear cells[2].

We report the observation of a GSSCM evolving in a small articulation (1st metatarsophalangeal joint of the left hallux), whose management consisted of an excision of the mass and a definitive arthrodesis given the extent of joint damages.

CASE REPORT

We report the case of a 32-year-old patient who, two years earlier, showed an initially painful gradual increase in the volume of his metatarsophalangeal articulation of the right hallux. The patient subsequently presented a blockage of his articulation and an embarrassment to the footwear. Physical examination revealed a firm mass, fixed to the deep planes without skin invasion (figure 1).



Figure 1: Clinical aspect

Standard X-ray and CT scan showed a tissue lesion with flaky periarticular circumferential ossifications of the first MTP with erosion of the head and neck of the 1st metatarsal and an articular pinching (figure 2).

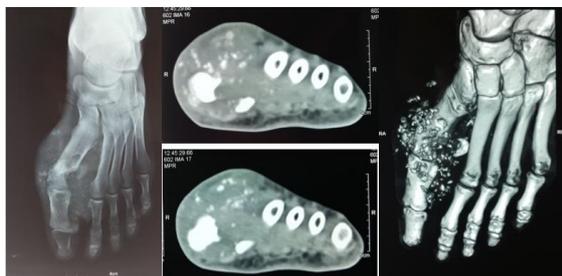


Figure 2: AP X-ray and CT-Scan aspects with 2D and 3D reconstruction

MRI revealed an aggressive-looking voluminous tumoral process of the soft tissues of the forefoot, seat of calcifications, associated with an abnormal bone signal and cortical notches of the first

metatarsal evoking chondrosarcoma or synovial tumor (figure 3).

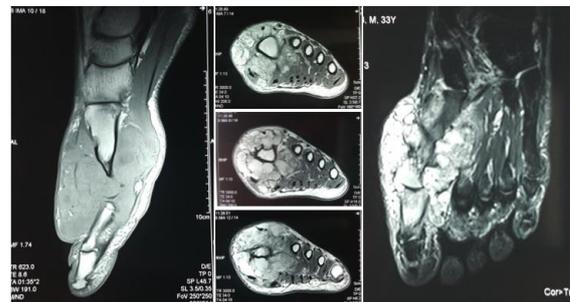


Figure 3: MRI aspect

In view of the volume of the tumor and its destructive appearance, a biopsy was performed confirming the diagnosis of synovial osteochondromatosis without associated cellular atypia. We performed an extensive synovectomy by a wide medial approach, taking away the entire pathological tissue (figure 4).



Figure 4: tumor resection

In view of the destabilization secondary to the wide excision and to the state of the articular cartilage, a final arthrodesis of the MTP was realized. The follow-up were simple with joint fusion, disappearance of pain and of the embarrassment to the footwear and absence of recurrence at the last recoil (20 months) (figure 5)



Figure 5: clinical aspect and X-ray control at the last follow up

DISCUSSION

Primary synovial osteochondromatosis (SOC) is characterized by a synovium cartilaginous metaplasia responsible for the sub-synovial production of multiple hyaline cartilage nodules. It is a benign neoplastic process that combines: clonal chromosomal abnormalities mainly chromosomes 5 and 6[3] and chromosome translocation 12-18[4], secretion of fibroblast growth factors and their receptors[3,5], a deregulation of differentiation of progenitor mesenchymal cells[6], an increase in collagen type II synthesis with an elevated concentration of the pro-collagen II C peptide marker[7].

It is a rare benign disorder (incidence 1: 100,000)[8], but often recurrent[5], which mostly affects the male sex (sex ratio: 2/1) between the 3rd and 5th decade[9]. It is also a primary condition that sometimes leads to joint destruction and osteoarthritis[10,11]. Mostly described in the large joints (knee, hip, elbow, ankle...)[12] rarely in the small joints (temporomandibular[13], metacarpophalangeal[14,15], interphalangeal[5], Talo-navicular[16], stero-clavicular [17], metatarso-phalangeal [18–20]...) where it causes osseous destruction and dislocations. The cartilaginous nodules may be free in the articular space or embedded in the synovium which can be thickened[8].

Usually the nodules are millimetric. However, in rare cases, the nodules cluster

together to form a large mass, or one of the nodules grows to a significant extent. Edeiken[1] gives the name of giant solitary synovial chondromatosis (GSSCM) to masses exceeding 1cm in diameter. Less than 20 cases of GSSCM are described in the Anglo-Saxon literature[1,2], whose sizes vary between 16 and 200 mm. The destructive and erosive character is in this situation more pronounced, especially when it comes to reaching a small joint like our patient. Destruction is equally relevant to epiphysis, metaphysis and structures of articular capsulo-ligamentary stabilization.

Histologically, the nodules show clusters of chondrocytes with large areas of chondroid matrix, without myxoid change. Chondrocytes show more or less marked cellular atypia and often have two nuclei. These facts should not lead to a false diagnosis of chondrosarcoma.

Synovial chondromatosis should not be confused with intra-articular loose bodies, also called secondary osteochondromatosis, which result from pre-existing joint damage, usually osteoarthritis. Synovial chondromatosis, which has evolved for a long time, can itself lead to an osteoarthritic model, and the two types of lesions are then entangled.

In its giant form, the differential diagnosis with chondrosarcoma must nonetheless be mentioned before the surgical management [2,21,22], especially since there are cases of chondrosarcoma occurring on joints which are subject to synovial chondromatosis without the causal relationship being established[22,23].

The management consists of excision of foreign bodies and synovectomy by arthroscopy in the usual forms and an open excision of the whole process in giant forms after biopsy confirming benignity. The management of joint wear is done on a case-by-case basis (abstention,

arthroplasty, arthrodesis) depending on the extent of the damage and the articulation involved [24].

CONCLUSION

Osteochondromatosis of small joints is infrequent and is exceptional in its giant form. In these latter a certain degree of atypia is sometimes found and is not systematically sign of malignancy. Synovial involvement is important because it is practically pathognomonic of this pathology and excludes differential diagnosis of chondroma or chondrosarcoma[25].

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